Visceral artery aneurysms in Von Recklinghausen's neurofibromatosis

Réda Hassen-Khodja, MD, Serge Declemy, MD, Michel Batt, MD, Jérôme Castanet, MD, Christophe Perrin, MD, Jean-Paul Ortonne, MD, and **Pierre Le Bas, MD,** *Nice, France*

We report the case of a patient with Von Recklinghausen's neurofibromatosis in whom two visceral artery aneurysms were diagnosed: a 4 cm aneurysm originating from the common hepatic artery and a smaller aneurysm originating from the superior mesenteric artery. The hepatic artery aneurysm underwent successful embolization. Because of the patient's poor general condition, the superior mesenteric aneurysm was considered inoperable and has been kept under surveillance by ultrasonography. Arterial involvement in Von Recklinghausen's neurofibromatosis is a well-known but infrequent occurrence. Stenotic lesions predominate, with the renal arteries being the site of predilection. Aneurysmal defects are less common, and involvement of the visceral arteries is exceptional. Only three reports of superior mesenteric artery aneurysm in patients with Von Recklinghausen's neurofibromatosis were found in the literature, and hepatic artery aneurysm has never previously been described in this disease. (J Vasc Surg 1997;25:572-5.)

Von Recklinghausen's neurofibromatosis, a phacomatosis inherited as an autosomal dominant trait of variable expressivity, occurs with an incidence of 1 per 3000.[1] Arterial involvement is a well-recognized but infrequent phenomenon.[2-6] Often multiple stenotic or aneurysmal arterial lesions can affect any of the arterial territories.[7] We report the rare case of a patient with Von Recklinghausen's neurofibromatosis in whom abdominal pain prompted diagnosis of two visceral artery aneurysms.

CASE REPORT

This 55-year-old woman was hospitalized in August 1993 for pain in the right lower quadrant. She had been monitored by our dermatology department for neurofibromatosis diagnosed on the basis of an association of multiple café au lait spots, neurofibromas, and Lisch nodules. This patient had had restrictive respiratory insufficiency for several years as a result of severe kyphoscoliosis caused by neurofibromatosis. Physical examination revealed a tender, pulsatile, space-occupying mass in the right quadrant. Abdominal computed tomography showed a large arterial aneurysm (Fig. 1). Aortography demonstrated that there were actually two saccular aneurysms. The largest measured 4 cm in diameter and was located 2 cm from the origin of the hepatic artery just upstream of the origin of the gastroduodenal artery (Fig. 2); the second aneurysm, which had developed on the superior mesenteric artery approximately 4 cm from its origin, was 1.5 cm in diameter (Fig. 3).

This patient was considered a poor candidate for surgical correction because of her severe respiratory insufficiency and chronic cor pulmonale, contraindicating laparotomy. Embolization of the hepatic artery aneurysm was performed in two stages at a 1-week interval because of the duration of the procedure, the quantity of contrast material required, and the embolization material used. Coil occlusion (Cook Co., Bloomington, Ind.) resulted in complete thrombosis of the aneurysm. The small size and location of the superior mesenteric artery aneurysm precluded operative intervention, and regular surveillance was initiated by ultrasonography and computed tomography. The patient is currently well after a 2-year follow-up.

DISCUSSION

Arterial vasculopathy in Von Recklinghausen's disease is a well-known but infrequent occurrence.[2-6] Most cases involve arterial stenoses in children or young adults. Renal localizations predominate.[2,6,8] Renovascular hypertension is the most frequent clinical manifestation in children. In adults arterial hypertension is often due to a pheochromocytoma associated with the neurofibromatosis.[9] More rarely, stenosis affects the intracranial arteries,[10-13] the supraaortic trunks,[11] the thoracic aorta,[14] the pulmonary artery,[11] or the visceral arteries.[11,15]

From Service de Chirurgie Vasculaire, Hôpital St. Roch, and Service de Dermatologie and Service de Pneumologie, Hôpital Pasteur.

Reprint requests: R. Hassen-Khodja, MD, Hôpital St. Roch, 5, rue Pierre Dévoluy, 06006 Nice, France.

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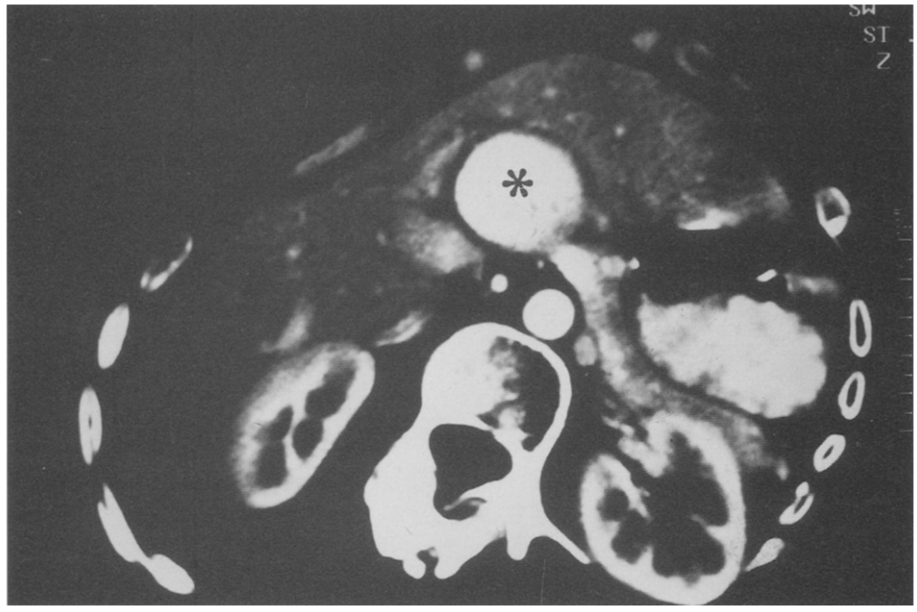

Fig. 1. Abdominal computed tomograph: large hepatic aneurysm *(star)*.

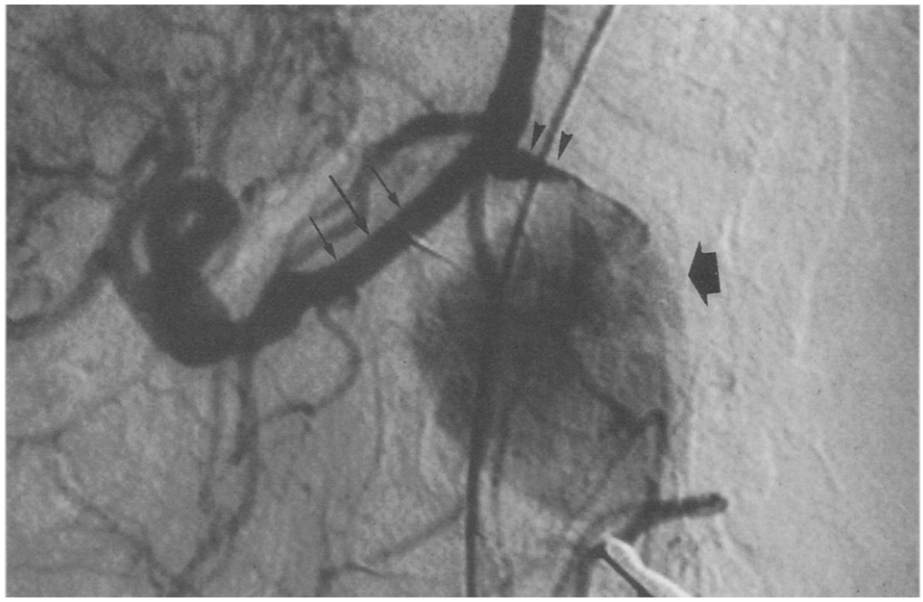

Fig. 2. Abdominal aortograph. Hepatic artery aneurysm *(large arrow);* common hepatic artery *(arrowheads);* splenic artery *(small arrows)*.

Aneurysmal defects are less frequent. Arterial vasculopathy in a patient with Von Recklinghausen's disease was first described in 1945 by Reubi.[16] These often multiple aneurysmal dilatations or associations of stenotic and aneurysmal lesions reflect the extensive nature of arterial involvement.[7] Aneurysms have been reported on the supraaortic trunks,[17,18] the descending thoracic aorta,[17] the renal arteries or their branches,[4,7,15,19] and the intracranial arteries.[3,9]

Aneurysms of the visceral arteries are exceptional; the major concern is the splenic artery.[4,7,15,20] Hepatic artery aneurysm has never previously been described in the English language literature. Only three cases of superior mesenteric artery aneurysm have

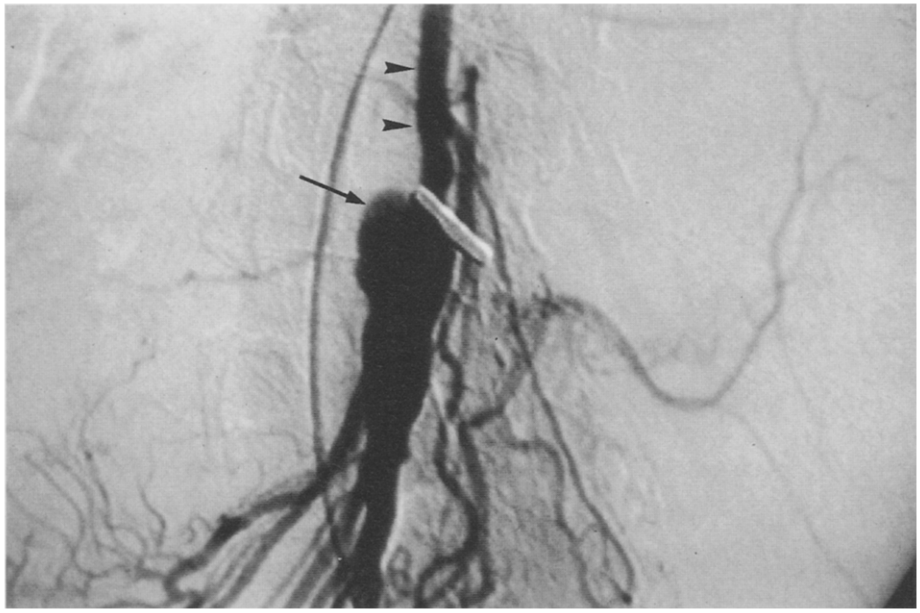

Fig. 3. Abdominal aortography. Superior mesenteric artery aneurysm *(arrow);* superior mesenteric artery *(arrowheads).*

been reported in Von Recklinghausen's neurofibromatosis.[4,15,21] Fye et al.[4] described multiple small aneurysms on the branches of the superior mesenteric artery. Zochodne[21] reported a 2 cm diameter aneurysm of the superior mesenteric artery associated with coarctation of the abdominal aorta and poststenotic changes in the renal arteries. Henley and Kaude[15] mentioned a superior mesenteric artery associated with multiple renal artery aneurysms but did not detail the case. Our patient had two aneurysmal defects, one on the common hepatic artery and another on the superior mesenteric artery.

The histopathologic features of arterial lesions in Von Recklinghausen's disease consist of fibrodysplastic changes in the media with segmental fragmentation of the vessel wall caused by atrophy of the muscularis and formation of saccular aneurysms. Both aneurysms in our patient were of the saccular type. Fusiform aneurysms are less common and are often poststenotic.[5,7,16,22,23] Because histologic confirmation of the nature of the visceral artery aneurysms was not obtained, their relationship to the patient's neurofibromatosis remains unproven, but it seems most probable.

The pathophysiologic characteristics of vascular lesions in neurofibromatosis remain controversial.[5,8,16,22] The alterations observed in the media and the adventitia appear compatible with the mesodermic and neuroectodermic tissue involvement in this disease.[5,7,16,22]

The risk of degeneration of these arterial aneurysms also remains unclear. Rupture, a potential complication,[4,24-26] appears rare. The prognosis for neurofibromatosis, which is unpredictable for a given subject, depends essentially on the existence of cerebral tumoral lesions, which are responsible for death in 72% of cases.[1]

Because our patient was considered a poor candidate for surgery, only the large painful hepatic artery aneurysm was managed by embolization. Regular surveillance of the superior mesenteric artery aneurysm by ultrasonography and computed tomography was initiated.

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